

Wernicke Encephalopathy: A Case Report

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Abstract

Wernicke encephalopathy is a medical emergency cause by thiamine or B1 deficiency that results life-threatening brain disruption, confusion, staggering and stumbling, lack of coordination, and abnormal involuntary eye movements. Chronic alcohol intake is the most common reason but it is also common in prolonged intravenous feeding, fasting, refeeding after starvation, prolonged vomiting such as hyperemesis gravidarum, anorexia nervosa, patients receiving dialysis, thyrotoxicosis, congestive heart failure with long term diuretic treatment. In non-alcoholic patients Wernicke Encephalopathy might develop due to the exclusion of upper portions of the gastrointestinal tract (e.g. after gastrectomy, gastrojejunostomy, gastric bypass surgery) or malabsorption. Other described conditions in which Wernicke Encephalopathy may develop include HIV/AIDS and several types of malignancy (inoperable gastric cancer, leukaemia and lymphoma).

Wernicke encephalopathy, if not recognized and treated, can become irreversible.

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I. Case Study

A 50-year-old man was admitted in our Hospital with the chief complaint of acute confusion, visual hallucination, persecutory idea, restlessness and disorientation and gait ataxia for 6 days . He had multiple episodes of vomiting 5 days back . He had weakness in lower limbs, inability to walk, ataxic gait and forgetfulness for last four months. He had been an alcoholic for 23 years and was still drinking alcohol at the time of this illness. During admission, vitals were stable with normal systemic examination. Physical examination showed a drowsy and disorientated man with a body temperature of 38.C He showed mild intentional tremor, symmetrical mild weakness, diminished deep tendon reflexes in all limbs, horizontal nystagmus, impaired finger-nose and heel-shin test, and dysdiadokinesia. Concentration was impaired. Disorientation to time and place was present. Memory was impaired. He also had positive signs of chronic liver disease. Other findings were unremarkable .

The complete blood count, serum glucose, BUN, creatinine, electrolytes, urinary analysis and chest X-ray were within normal limits. Liver function test revealed an elevated Liver enzyme level. The MRI of the brain showed multiple,scattered, small non-enhanced low signal intensity on T1WI, high signal intensity on T2WI involving at bilateral basal ganglias, thalami, midbrain, pons, and periventricular regions with evidence of brain atrophy. The mamillary bodies could not be identified. A cerebrospinal fluid analysis was within normal limits. He was treated with parenteral thiamine intravenously 500 mg in 0.9% normal saline thrice daily for two consecutive days followed by 500 mg intravenously once daily for an additional 5 days. The nystagmus, double-vision, incoordination, weakness of lower limbs, ataxia improved but memory problems, and loss of insight remained.

II. Discussion

Wernicke Encephalopathy is a medical emergency caused by thiamine or B1 deficiency. This micronutrient, a water-soluble vitamin, is absorbed primarily in the duodenum, acts as a co-factor in carbohydrate metabolism, and is important in neuron cell function. The human body cannot synthesize thiamin and regular dietary intake of thiamin is essential. The most common cause is found in alcoholism but it is also common in prolonged intravenous feeding, fasting, refeeding after starvation, prolonged vomiting such as hyperemesis gravidarum, anorexia nervosa, patients receiving dialysis, thyrotoxicosis, congestive heart failure with long term diuretic treatment. However, it is also caused by a genetic abnormality resulting from a defecation in transketolase structure that diminished binding with coenzyme thiamine pyrophosphate. Thiamin is a cofactor for transketolase, alpha-ketoglutarate dehydrogenase, pyruvate dehydrogenase, and branch-chain alpha-keto acid dehydrogenase that are

essential in intermediate carbohydrate metabolism. A decrease in their activity may lead to increased buildup of toxic intermediates. The buildup of intermediates that are unable to be metabolised may induce damage in areas of the brain where excess accumulation occurs.

Thiamine may also function in axonal conduction and synaptic transmission. Thiamine deficiency causes alteration of cerebral energy mechanisms, diminishes nerve impulse transmission at synapses and impairs DNA synthesis that characterized by necrosis of

nerve cells and myelinated structures. Wernicke Encephalopathy may evolve as minor episodes comprising combinations of these more subtle features and that the clinical form of the disease develops after repeated episodes of subclinical encephalopathy, which is supported by autopsy findings. The characteristic brain lesions include symmetrical discoloration of structures surrounding the third ventricle, aqueduct, and fourth ventricle, with petechial hemorrhages in occasional acute cases and atrophy of the mamillary bodies in a majority of chronic cases. Involvement of the third and sixth nuclei and tegmentum account for the ophthalmoplegias.

Wernicke Encephalopathy has an acute onset characterised by nystagmus, ophthalmoplegia, mental status changes and unsteadiness of stance and gait, although this triad is seen only in 16% of patients. About 82% of patients have mental status changes according to autopsy-based series. These changes occur due to involvement of thalamic or mammillary bodies and range from acute confusional state to mental sluggishness, apathy and inability to concentrate, and, if not treated, it leads to coma and death. The common ocular abnormalities are

Horizontal nystagmus and lateral rectus muscle palsies. The other findings are conjugate-gaze palsies, ptosis, pupillary abnormalities, complete ophthalmoplegia, and papilledema. It can be one-sided, or on both sides. The loss of equilibrium seen in the early stages is due to vestibular paresis while the wide-based ataxic gait seen in the subacute and chronic phases is due to cerebellar dysfunction. Diagnosis of Wernicke Encephalopathy has been based on history, clinical symptoms and the laboratory investigations such as low serum thiamine and transketolase activity and elevation of pyruvate level.

MRI is currently considered the most valuable method to confirm a diagnosis of WE. MRI shows symmetric involvement of the mammillary bodies, the tectal plate, the periaqueductal grey matter and the periventricular region of the third ventricle including the paramedian thalamic nuclei. Signal hyperintensities on T2-weighted sequences, FLAIR and diffusion-weighted images within the posteromedial thalami and surrounding the third ventricle are the most common abnormality described in patients with Wernicke Encephalopathy. The CT scan of the brain occasionally shows low attenuation with or without enhancement in the involved regions, especially with symmetric low density in the thalami.

The mortality rate is 10-20% and the complications are permanent loss of memory and cognitive skills. The treatment may correct all abnormalities so it must be viewed as a medical emergency. The goal of treatment is to control symptoms as much as possible and to prevent progression of the disorder. Thiamine is the drug of choice for treatment of Wernicke Encephalopathy. Patients with suspected Wernicke Encephalopathy require immediate parenteral administration of thiamine. A recommended regimen is 500 mg of thiamine intravenously, infused over 30 min, three times daily for two consecutive days and 500 mg intravenously once daily for an additional 5 days. To conclude, acute Wernicke Encephalopathy continues to be a rare but life-threatening condition often overlooked in the non-alcoholic population, resulting in the further progression of an easily treatable condition. Treatment should be initiated at the earliest possible time to avoid persistent brain damage. The prognosis of Wernicke Encephalopathy is favorable if diagnosed and treated early.

Reference

- [1]. Victor M, Adams RD, Collins GH. The Wernicke-Korsakoff syndrome and related neurologic disorders due to alcoholism and malnutrition 2nd edn Philadelphia, PA: FA Davis, 1989
- [2]. Victor M. The Wernicke-Korsakoff syndrome. In: Vinken PJ, Bruyn GW. eds. Handbook of clinical neurology. vol 28, part II Amsterdam: North-Holland; 1976:243–70
- [3]. Harper CG, Giles M, Finlay-Jones R. Clinical signs in the Wernicke-Korsakoff complex: a retrospective analysis of 131 cases diagnosed at necropsy. *J Neurol Neurosurg Psychiatry* 1986;49:341–5
- [4]. Jung ES, Kwon O, Lee SH, et al. Wernicke's encephalopathy in advanced gastric cancer. *Cancer Res Treat* 2010;42:77–81
- [5]. Merkin-Zaborsky H, Ifergane G, Frisher S, et al. Thiamine-responsive acute neurological disorders in nonalcoholic patients. *Eur Neurol* 2001;45:34–7
- [6]. Dreyfus PM. Clinical application of blood transketolase determinations. *N Engl J Med* 267:596–
- [7]. Weidauer S, Nichtweiss M, Lanfermann H, et al. Wernicke encephalopathy: 2003; 2003;13:1001–9
- [8]. Unlu E, Cakir B, Asil T. MRI findings of Wernicke encephalopathy due hunger strike. *Eur Radiol* 2006;57:43–53
- [9]. Pandit L. Differential diagnosis of white matter diseases in the tropics: an overview. *Indian Acad Neurol* 2009;12:12–21
- [10]. Cook CC, Hallwood PM, Thomson AD. B vitamin deficiency and neuropsychiatric syndromes in alcohol misuse. *Alcohol* 1998;33:317–36
- [11]. Blansjaar BA. Korsakoff minus wernicke syndrome. *Alcohol and Alcoholism*. 1992; 27(4):
- [12]. Bonucchi J, Hassan I, Policeni B, Kaboli P. Thyrotoxicosis associated Wernicke's encephalopathy. *Journal of General Internal Medicine*. 2008; 23(1): 106-9.

- [13]. Caine D, Halliday GM, Kril JJ, Harper CG, Alfred RP. Operational criteria for the classification of chronic alcoholics: identification of Wernicke's encephalopathy. *Journal of Neurology, Neurosurgery, and Psychiatry*. 1997; 62: 51-60.

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